Picture Stories

Rowell syndrome in a nine year old child

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Introduction

Rowell syndrome (RS) in 1963 first described the co-existence of lupus erythematosus (LE) and erythema multiforme (EM) with positive rheumatoid antigen (RA), speckled antinuclear antigen (ANA) and precipitating antibody to saline extract of human tissue (anti-SjT)[1]. Anti-SjT is now regarded as anti-Ro antibody. Subsequently diagnostic criteria were established that all the major criteria and at least one minor criterion should be present so as to be called RS. The major criteria are presence of LE, EM and positive speckled ANA. The minor criteria are chilblains, positive RA and positive anti-Ro or anti-La[2].

Case report

A nine year old girl was admitted with fever for two weeks with fatigue and generalised muscle pain for the same duration. There were also complaints of a prominent painless oral ulcer and multiple reddish lesions on the face, back and extremities for the last one week. There was no history of upper respiratory tract infection or any drug intake before this episode. On examination, there were multiple, hyperpigmented, erythematous, well-defined and confluent papules and plaques distributed symmetrically over the face, back and extremities, including the palms and soles. Some of the lesions were "targetoid" in appearance. Lesions over face were approximately 1 cm x 1 cm in size, erythematous in colour, with a lighter periphery and deeper dusky centre. They affected mostly the cheeks in almost a bilaterally symmetrical manner. Some of the bigger plaques on the face later became scaly. There was a well-defined ulceration on the surface of the hard palate and prominent crusting on the lips. (Figure 1).

General examination revealed a moderate degree of pallor but the vital parameters were normal. Remaining systemic examination revealed no abnormality. Blood tests revealed pancytopenia (haemoglobin 9 g/dl, white cell count 2800/cu mm, platelet 138,000/cu mm) with normal liver and renal functions. Urine examination did not reveal any significant proteinuria or casts. Tests for malaria, dengue, typhus, enteric fever, blood culture and urine culture were negative. Upon the suspicion of co-existence of systemic lupus erythematosus (SLE) and EM, subsequent tests were performed. They showed positive ANA >640, speckled pattern, low C3 and C4 levels, positive anti ds DNA, negative rheumatoid factor and positive anti-Ro/ anti-La antibody. Chest X-Ray, echocardiography, ultrasonography of abdomen and ophthalmological examination were normal. Skin biopsy revealed hyperkeratosis, necrotic keratinocytes, epidermal necrosis, vacuolar degeneration of the dermal-epidermal junction, perivascular lymphocytic infiltrate and papillary dermal oedema, suggestive of EM. A diagnosis of Rowell syndrome was made. She had received a course of empirical therapy with intravenous acyclovir and ceftriaxone for the last seven days, with no improvement of symptoms or signs. Subsequently we started prednisolone 2 mg/kg/day in three divided doses and hydroxy-chloroquine. Child improved gradually over the next one month. No fresh lesion appeared during follow-up and hence steroids were being tapered slowly.

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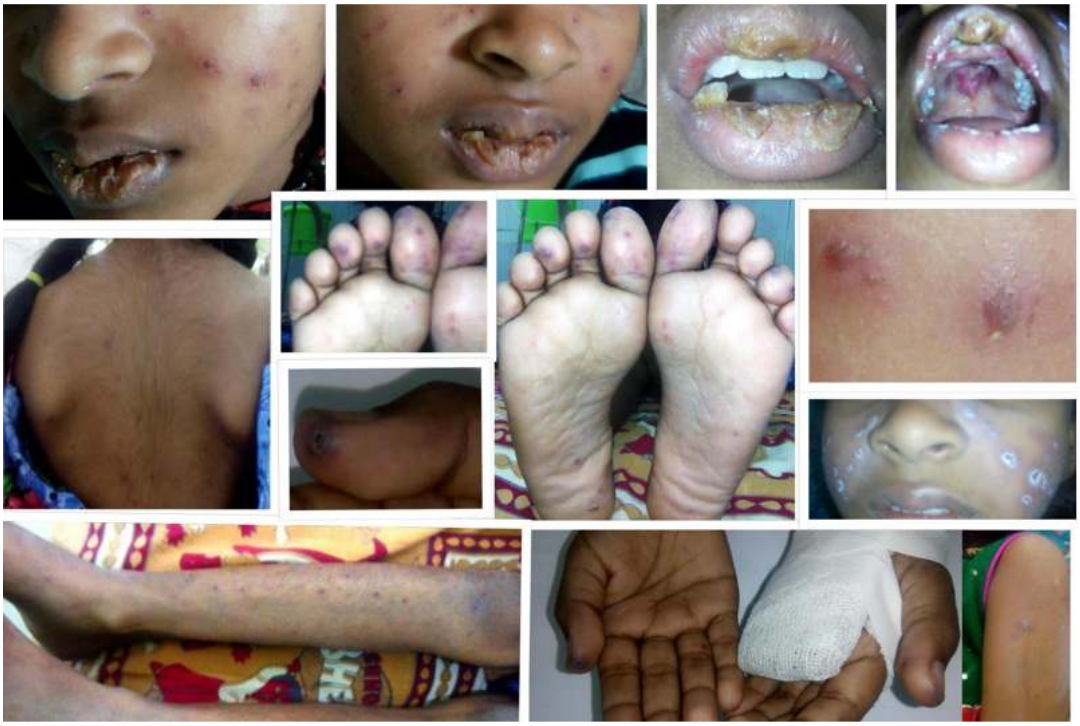

Figure 1: ***Various skin and mucous membrane lesions as seen in this child***

Discussion

SLE was diagnosed in this child by the presence of oral ulcers, pancytopenia, positive ANA, and anti ds-DNA antibodies[3]. Prolonged fever, fatigue, myalgia and female sex (females are 9 times more affected than males) were the other clues for SLE. EM was suspected by the typical morphology of the rash and confirmed on biopsy. However, there were many differential diagnoses for EM. For example, urticarial rash tends to be transient, Stevens Johnson syndrome and fixed drug eruptions may have a drug history, while there are tense bullae in bullous pemphigoid. Severe mucosal involvement is seen in Stevens Johnson syndrome and paraneoplastic pemphigus, while there is no mucosal involvement in Sweet's syndrome. There should be typical palpable purpura in cutaneous small vessel vasculitis[4]. The two most common causes of EM are HSV infection and drug-intake. While the oral ulcers are always painful in herpetic gingiva-stomatitis, they are painless in as much as 82% of cases in SLE[5]. Besides, RS must be suspected when there is a long standing EM or when there is no response with acyclovir[6]. Hence, as there was co-existence of SLE and EM, with positive ANA and anti Ro/ anti-La antibodies, she was diagnosed as a case of RS according to the diagnostic criteria[2].

It should be noted that there are ongoing debates on whether RS is an overlap syndrome, a real association, or coincidence of DLE and EM. Some authors suggest RS to be a sub-entity of subacute LE with EM while others suggest it to be a different variant of cutaneous LE, a subtype of chronic LE, an independent LE subtype, or an autonomous type of cutaneous LE[2]. Tiwary *et al* took skin biopsy from both the targetoid lesions and the plaques in a RS patient and showed them to be histo-pathologically similar[7].

Besides, RS has been mostly reported in middle aged and elderly women (31 to 72 years)[(7)]. This case shows that RS should also be suspected in patients as young as nine years. To our best knowledge, ours is one of the youngest RS patients to be reported in the literature. The co-existence of EM in a paediatric patient showing few clinical signs of SLE should not be a distracting factor for the clinician and turn his attention from sending further investigations for lupus.

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